STAPHYLOCOCCAL PURPURA FULMINANS

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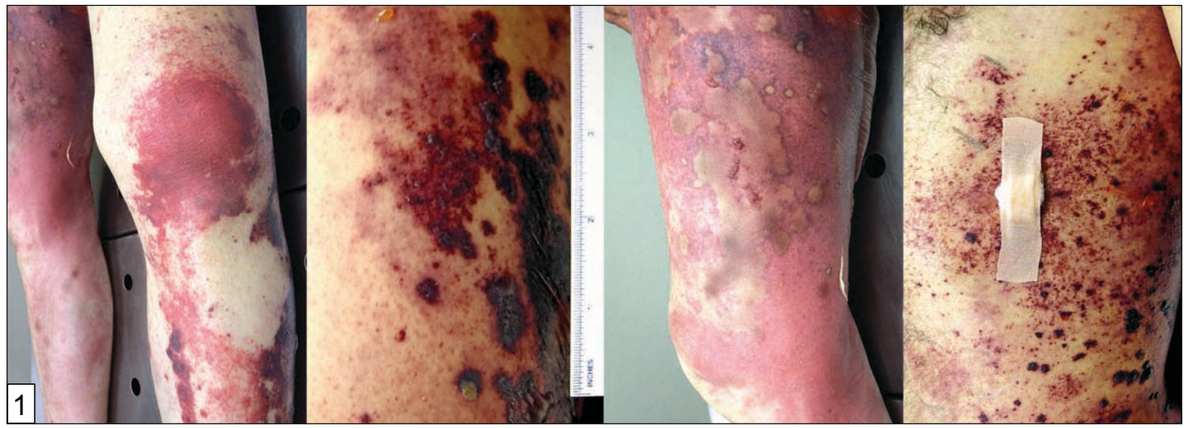
ABSTRACT

Purpura fulminans (PF) is associated with several infections; however, there are few reports of this entity in association with TSST-1 producing S. aureus. We report a 53-year-old male who presented with fever, progressive hemodynamic instability, multi-organ failure, and thrombocytopenia following lobectomy for a solitary lung metastasis of rectal adenocarcinoma. Additionally, he developed progressive generalized eruption of non-blanching purple to black macules, papules, and plaques on the trunk and extremities consistent with PF. He expired on post-admission day three. Autopsy examination revealed pleural purulent exudate, which grew toxic shock syndrome toxin-1 (TSST-1) producing Staphylococcus aureus. Pre-mortem and autopsy skin biopsies demonstrated subepidermal bullae, epidermal necrosis, and fibrin deposition within small cutaneous vessels with minimal lymphocytic infiltration. Vasculitis was not present. TSS associated PF may be highly under-recognized and much more common than reflected by the literature.

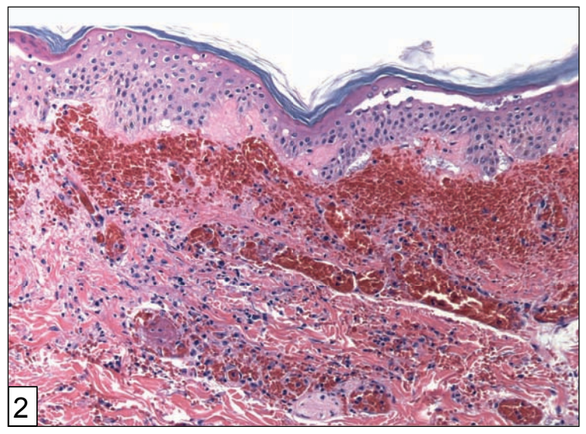
INTRODUCTION

PF is a life threatening condition characterized by hemorrhagic infarction of the skin due to disseminated intravascular coagulation and dermal vascular thrombosis. PF can be identified in three different conditions as follows: 1) “Neonatal PF,” which is associated with a hereditary deficiency of the anticoagulants Protein C and Protein S and Antithrombin III; 2)“Idiopathic PF,” which is associated with an initiating febrile illness, however deficiency of Protein S is considered to be central to the pathogenesis; and 3) the most common type, “Sepsis-associated PF,” which is not specific for any infection, but is commonly associated with meningococcemia (10-20% of meningococcemia cases) and less frequently with groups A and B ß-hemolytic streptococci, S. pneumonia, Haemophilus influenza. Sepsis-associated PF has four primary features: large, purpuric skin lesions; fever; hypotension; and disseminated intravascular coagulation. It has a high mortality rate and is commonly referred as staphylococcal toxic shock syndrome.

SKIN GROSS EXAMINATION



HISTOPATHOLOGICAL EVALUATION



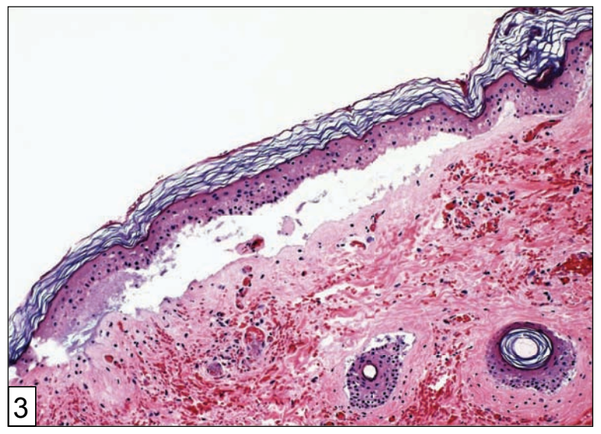


Figure 1. The entire body was covered with diffuse, bright red papules and plaques (2 mm to 3 cm). Within two days these areas became flat and dark purple and began to slough. **Figure 2.** Pre-mortem skin biopsy (Day 2 post-admission) demonstrated extensive hemorrhage in papillary dermis, subcorneal splitting, thrombi in small vessel and mild peri-vascular infiltrate. No vasculitis was present. **Figure 3.** Postmortem skin biopsy (Day 3 post-admission) showed similar findings which pro-gressed to form subepidermal bullae and epidermal necrosis. These changes are consistent with disseminated intravascular coagulation (DIC) and purpura fulminant.

CONCLUSION

Purpura fulminans (PF) is an acute illness and is typically characterized by disseminated intravascular coagulation (DIC) and purpuric skin lesions. Usually PF is synonymous with severe meningococcemia, however, meningococcal infections are relatively rare. S. aureus bacteremia, on the other hand, occurs much more frequently than does meningococcemia, but because it is not categorized by the Centers for Disease Control and Prevention as a notifiable disease, precise data on the incidence are not available. Previously, S. aureus infections have only rarely been complicated by purpura fulminans. Kravitz et al. reported the first 5 cases of TSS associated purpura fulminans due to Staphylococcus aureus strains that produced high levels of the superantigens toxic shock syndrome toxin–1 (TSST-1), staphylococcal enterotoxin serotype B (SEB), or staphylococcal enterotoxin serotype C (SEC). Staphyloccocal toxic shock syndrome is a rapidly progressive, serious systemic illness caused by superantigens produced by S. aureus and can be associated with any staphylococcal infection. Staphylococci are rarely cultured from blood, but can be cultured from mucosal sites or localized abscesses. TSS associated PF may be highly under-recognized and much more common than reflected by the literature.

CASE REPORT

The patient was a 53-year-old-male with a history of moderately differentiated adenocarcinoma of rectum, status post chemo/radiotherapy. Following detection of metastatic rectal adenocarcinoma (solitary nodule of 2.7 cm), he under-went a right lung middle lobectomy. Due to a persistent air leak, he was discharged with a right chest tube in place. Seven days later, he was brought to emergency department (ED) in shock, febrile and unresponsive. Several red and/or purple rash in groin area were discovered on admission. He rapidly decompensated and was intubated and admitted in ICU and was put on vasopressive and broad spectrum antibiotics. He developed acute renal failure and lactic acidosis, showed signs of shock liver, profound coagulopathy and thrombocytopenia. On hospital day 2, he began developing diffuse, bright red papules and plaques (2 mm to 3 cm) covering his entire body including his testis and scrotum. These areas then became flat and dark purple and began to slough. His skin lesions were consistent with purpura fulminans (**Figure 1**). Laboratory data showed marked leukocytosis and left shift suggesting an infection, however all cultures were negative until that point. Due to severely deteriorating condition of the patient, massive tissue destruction and shock, in the setting of stage 4 cancer, the family opted for comfort care and the patient passed away. At autopsy, gross examination of the lungs revealed yellow gelatinous and fibrous exudates overlying the surface of both lungs. Cultures of pleural fluid and the exudates grew staphylococcus aureus. In addition, assay of these cultures showed TSS Toxin 1 (TSST-1) producing Staph Aureus. Pre-mortem and autopsy skin biopsies demonstrated progression of subcorneal to subepidermal bullae respectively, epidermal necrosis, and fibrin deposition within small cutaneous vessels with minimal lymphocytic infiltration. Vasculitis was not present.

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